

Case Report

A case of autoimmune induced necrotizing myopathy

Jaison Udani, Kirill Alekseyev*, Malcolm Lakdawala, Jitendra Patel, Marc Ross

Department of Physical Medicine and Rehabilitation, Kingsbrook Jewish Medical Center, Brooklyn, NY, USA

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***Correspondence:**

Dr. Kirill Alekseyev,

E-mail: kirill.alekseyev@gmail.com

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ABSTRACT

A 31-year-old female, with no previous past medical history, presented to the emergency department (ED) two weeks after falling while moving furniture complaining of increasing weakness and swelling. Initially, she had minor ankle pain after falling but this progressed to bilateral ankle pain and eventually bilateral shoulder pain. Creatine Kinase (CK) levels were drawn and results showed a level of 30,186. She was diagnosed with rhabdomyolysis and aggressive IV hydration was initiated. After 3 weeks, her CK levels did not decrease below 15,000. A muscle biopsy was performed and revealed a necrotizing myopathy with high probability of autoimmune etiology.

Keywords: Autoimmune, Necrotizing myopathy, Rhabdomyolysis, Creatine kinase, Debility, Rehabilitation

INTRODUCTION

Necrotizing autoimmune myopathy (NAM) is a relatively newly recognized subgroup of idiopathic inflammatory myopathies, which despite diverse causes, have the common histopathological features of myocyte necrosis without significant inflammation along with simultaneous muscle fiber regeneration.¹ Patients present with a sub-acute severe symmetrical proximal myopathy, associated with a markedly elevated creatine kinase level. These are most likely immune-mediated, as they respond to immunotherapy.² Given the fact that the condition can respond to immunosuppressive therapies, further supports that this condition is immune-mediated, although there is a lack of substantial immune cell invasion of muscle.¹ This uncommon condition may be confused with muscle damaging disease processes such as polymyositis (PM) or rhabdomyolysis.³ Medication induced necrotizing myopathy should also be ruled out especially with the prolonged use of statins. In patients with auto-immune necrotizing myopathy, which is sometimes classified with PM, scattered necrotic muscle fibers are present.

Necrotizing myopathy is sometimes associated with anti-signal recognition particle (SRP) antibodies. The small amounts of immune cells that are present in muscle appear to be macrophages that are invading myofibers secondary to necrosis of these myofibers. There is no evidence of invasion of myofibers that appear otherwise morphologically normal, and inflammatory cells are not present in perivascular or endomysial locations, unlike in dermatomyositis (DM), PM, or inclusion body myositis (IBM). Unlike those with DM, these patients do not exhibit peri-fascicular atrophy. Sometimes inflammatory cells are found around small blood vessels, and thickened basement membranes are seen. One hypothesis is that circulating soluble molecules may injure myofibers, but little is known regarding the mechanisms underlying this entity.^{4,5}

CASE REPORT

We present a case of a 31 year-old female that presented with generalized weakness. The patient stated she had fallen walking up stairs 3 weeks ago while moving furniture. She experienced bilateral ankle swelling

following the fall. Patient stated that the bilateral ankle swelling progressed to bilateral lower extremity pain along with new onset bilateral shoulder pain which prompted her to go to her primary medical doctor. She was prescribed a muscle relaxant and ibuprofen which did not relieve the pain. After a week her symptoms did not improve and she went to the emergency department. There they increased her medications with the addition of tramadol. Later that week, she stated her heart was racing and felt dizzy which prompted her to go to the ED again. There she was diagnosed with rhabdomyolysis with a CK level of 30186. On physical examination; strength in bilateral upper extremities was 2/5 and bilateral lower extremities 2/5 with the exception of bilateral knee flexors and extensors at 4/5. Deep tendon reflexes were 0/4 throughout and limited by generalized edema. The patient was started on aggressive IV hydration for rhabdomyolysis. Her CK levels remained elevated above 15,000 for 3 weeks. During this time, she was transferred to the medicine service for further evaluation of elevated CK levels and a muscle biopsy was performed. The report of the biopsy stated: vigorous turnover (myonecrosis/regeneration) of muscle fibers is consistent with clinically reported rhabdomyolysis, but there are chronic aspects in the pathology of the muscle. Small collections of chronic inflammatory cells in the background of extensive myonecrosis/ regeneration, which include a sizable number of CD138(+) plasma cells, CD69(+) macrophages/histiocytes, CD3(+) T-cells, and CD20(+) B-cells, which raises a question of immune mediated disease. There were also reticulo-tubular aggregates (RTA's) amongst the endothelial cells of intramuscular microvessels. RTA's are peculiar endothelial abnormalities that can be associated with systemic lupus erythematosus, dermatomyositis, Sjogren's syndrome, mixed connective tissue disease, as well as HIV. Immunological testing for all of these disease processes were unremarkable. The overall morphological observation raised high suspicion for immune mediated etiology underlying this severe and active necrotizing myopathy. She was started on prednisone and the following day her CK level decreased by half from 14584 to 7791.

DISCUSSION

There are subtle differences in the clinical manifestations of the various metabolic myopathies, but one of these conditions should be suspected when the following clinical circumstances are present. Recurrent episodes of rhabdomyolysis after exertion or in association with fasting or a viral illness. The last two associations occur most commonly with carnitine palmitoyltransferase deficiency and other disorders of lipid metabolism. There is a history of exercise intolerance, recurrent cramps, and fatigue beginning in childhood, and episodes of pigmenturia occurring in adolescence. There is a family history of rhabdomyolysis or exercise intolerance, particularly in siblings, thereby suggestive of an autosomal recessive inheritance pattern. The individual

has normal strength and muscle enzymes during interictal periods. One exception is muscle phosphorylase deficiency, a disorder in which chronic muscle weakness may develop after repeated episodes and CK levels do not return to normal between attacks.⁶ Symptoms include an asymmetric or distal presentation of weakness, intermittent symptoms, painful muscles, marked muscle atrophy, family history of muscle disease, a history of medication that could be associated with myopathy and neuropathic symptoms or findings.^{6,7}

CONCLUSION

This is a case of a patient with autoimmune induced necrotizing myopathy with elevated levels of creatine kinase (CK) after a minor fall at home while moving furniture. The CK levels did not improve with aggressive IV hydration and the original diagnosis of rhabdomyolysis was excluded. A muscle biopsy was done and raised high suspicion of immune mediated etiology underlying this severe and active necrotizing myopathy. These patients can be distinguished from patients with rhabdomyolysis, PM, or DM by the persistence of symptoms and findings, including the elevation in CK in the absence of treatment with immuno-suppressives, dermatologic manifestations, and by their histopathologic changes. It may also be seen in association with an underlying autoimmune rheumatic disease, such as scleroderma or mixed connective tissue disease, as well as paraneoplastic syndrome or may be idiopathic. Patients originally diagnosed with rhabdomyolysis should be considered for this rare diagnosis when elevated CK levels do not return to normal after proper management and exclusion of other muscle damaging disease processes.

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